JACC Vol. 6, No. 2
August 1985:388–93

Two-Dimensional Echocardiographic Evaluation of Right Ventricular Size and Function in Newborns With Severe Right Ventricular Outflow Tract Obstruction

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Critical pulmonary stenosis or atresia with intact ventricular septum represents a congenital cardiac lesion for which the long-term prognosis appears to depend partly on the size of the right ventricle. Thus, the capability of noninvasive assessment of right ventricular size to predict operative outcome was examined in 15 infants (aged 1 to 30 days, mean 5.6) with severe right ventricular outflow tract obstruction (either critical pulmonary stenosis [7 patients] or pulmonary atresia with intact ventricular septum [8 patients]). Using echocardiography in two orthogonal subxiphoid views, right ventricular volume, wall thickness, area change fraction, ejection fraction and tricuspid anulus dimension were measured.

All patients with a normalized right ventricular end-diastolic volume of less than 5 ml/m^2 and a normalized tricuspid anulus dimension of less than 1.0 cm/m$^{2/3}$ required a shunt operation. Only one patient with a volume of more than 6 ml/m^2 and a normalized tricuspid anulus dimension of more than 1.4 cm/m$^{2/3}$ required more than relief of right ventricular outflow tract obstruction. In this patient, residual severe pulmonary stenosis necessitated the shunt procedure. One patient with a volume of more than 6 ml/m^2 had an anulus diameter of less than 1.4 cm/m$^{2/3}$ and one patient with an anulus diameter of more than 1.4 cm/m$^{2/3}$ had a volume of less than 6 ml/m^2; both required shunt procedures. It therefore appears that if either the ventricular volume or tricuspid anulus size is excessively small, a shunt procedure is necessary. Wall thickness, area change fraction and ejection fraction measurements were not significantly correlated with right ventricular volume or postoperative outcome.

The noninvasive (echocardiographic) assessment of right ventricular volume and tricuspid valve anulus dimension permits excellent prediction of postoperative outcome. In patients with severe right ventricular hypoplasia, the need for alternative surgical therapy may thereby be recognized before initial intervention.

(J Am Coll Cardiol 1985;6:388–93)

In newborns with severe right ventricular outflow tract obstruction (pulmonary atresia with intact ventricular septum or critical pulmonary stenosis), the pulmonary blood supply is dependent on the patency of the ductus arteriosus or, rarely, a systemic to pulmonary collateral vessel. With postnatal spontaneous closure of the ductus arteriosus, profound hypoxemia occurs. Although infusion of prostaglandin E_1 is an effective temporizing maneuver (1–12), surgical intervention is essential for survival. Nonetheless, a high mortality rate is associated with this lesion (13). Classification of pulmonary atresia with intact ventricular septum has been based on the size of the right ventricle (14–17), the size or configuration of the tricuspid valve (14,18) or additional right ventricular morphologic and functional findings (19). This angiographic classification has been used as a means of optimizing surgical intervention.

There have been few echocardiographic reports of the M-mode (20) and two-dimensional findings (21) in severe right ventricular outflow tract obstruction with intact ventricular septum, and none has attempted to assess indexes predictive of late outcome. It is the purpose of this report to define morphologic or functional echocardiographic variables that could predict the optimal surgical approach in these critically ill infants.

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0735-1097/85/$3.30

Table 1. Clinical Data in 15 Patients

Case	Age (days)	BSA (m^2)	Diagnosis		Prostaglandin Therapy	Operation	
			PDA	Other		First	Second
1	1	0.21	+	PS	+	V	–
2	2	0.22	+	PS	+	V	S
3	30	0.20		PS		V	–
4	1	0.17	+	PA/IVS	+	V+S	–
5	1	0.22	+	PA/IVS	+	V+S	–
6	30	0.25		PS		V	–
7	2	0.20	+	PA/IVS	+	V+F	S
8	1	0.18	+	PA/IVS	+	V	S
9	10	0.22	+	PS	+	V	–
10	1	0.24		PS		V	–
11	1	0.24	+	PA/IVS	+	V+S	–
12	1	0.18	+	PA/IVS	+	V	–
13	1	0.22	+	PA/IVS	+	V	–
14	1	0.18	+	PS	+	V+S	–
15	1	0.23	+	PA/IVS	+	V+F	–

BSA = body surface area; F = formalinization; PA/IVS = pulmonary atresia with intact ventricular septum; PDA = persistent ductus arteriosus; PS = pulmonary stenosis; S = Blalock-Taussig shunt; V = valvotomy. + = present; – = not performed.

Methods

Subjects. Between February 1983 and July 1984, 15 preoperative infants (age 1 to 30 days, mean 5.6; body surface area 0.17 to 0.25 m^2, mean 0.21) with severe right ventricular outflow obstruction underwent two-dimensional echocardiographic examination. Clinical and operative data are shown in Table 1. Eight patients had pulmonary atresia with intact ventricular septum and seven had critical pulmonary stenosis. The diagnosis of pulmonary atresia or pulmonary stenosis was documented by angiography and confirmed at surgery in each case. The right ventricular pressure was systemic or suprasystemic in all patients. All except three patients with severe pulmonary stenosis were treated with prostaglandin E_1 preoperatively. Six patients underwent a transpulmonary valvotomy combined with either a Blalock-Taussig shunt (four patients) or formalinization of the ductus arteriosus (two patients). In one of the latter two patients, a systemic to pulmonary shunt operation was subsequently performed. Two patients initially underwent only pulmonary valvotomy but subsequently required a systemic to pulmonary shunt procedure because of severe persistent cyanosis. In seven patients only a pulmonary valvotomy was performed. The echocardiographic data were not used to determine the selection of surgical procedure.

Echocardiography. The two-dimensional echocardiograms were performed with an ATL Mark 600 or a Diasonics CV 100 using a 5 MHz transducer. The right ventricular diaphragmatic and septal wall thicknesses were measured in anteriorly angled subxiphoid long- and short-axis views through the infundibulum (Fig. 1). The maximal dimension of the tricuspid valve anulus was measured in both views, averaged and adjusted for the cube root of body surface area. With the use of a Franklin Quantic 1200 echocardiographic review station, the endocardial border of the right ventricle was digitized at end-diastole and end-systole and the respective cross-sectional areas were calculated. Area change fraction was calculated for both planes as: (end-diastolic area – end-systolic area)/end-diastolic area. The total area change fraction was then derived as the average of long- and short-axis values. Applying a modified Simpson's rule, right ventricular volumes were calculated and adjusted for body surface area. Ejection fraction was calculated as: (end-diastolic volume – end-systolic volume)/end-diastolic volume.

Statistics. The relation between adjusted tricuspid valve anulus dimension and adjusted right ventricular end-diastolic volume was determined using linear and nonlinear regression analysis. Values for right ventricular wall thickness and tricuspid anulus dimension in both planes were compared using the *t* test for paired data. Linear regression analysis was applied to determine the relation between right ventricular end-diastolic volume and ejection fraction, area change fraction and right ventricular wall thickness and ejection fraction and area change fraction. A p value of 0.05 or less was considered statistically significant.

Results

Individual values for wall thickness, tricuspid valve anulus diameter, area change fraction, volumes and ejection fraction are shown in Table 2.

Right ventricular volume. All five patients with right ventricular end-diastolic volume of less than 5 ml/m^2 underwent a shunt operation as part of the primary procedure;

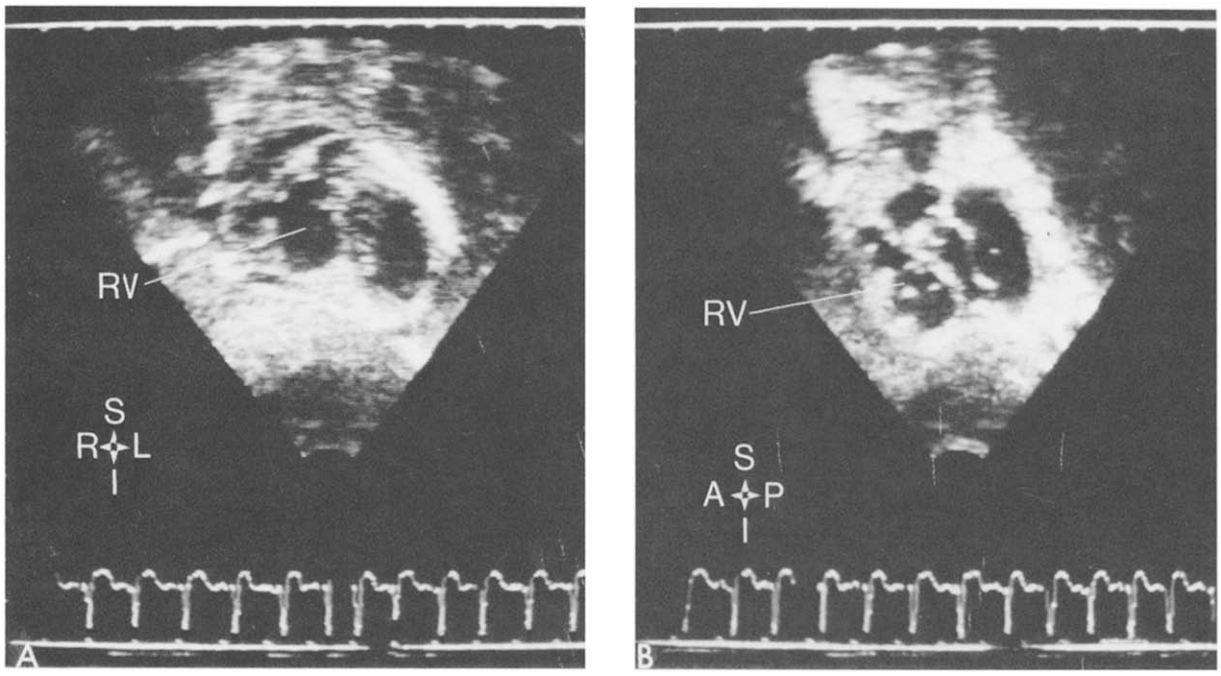

Figure 1. Long- **(A)** and short- **(B)** axis views of the right ventricle (RV) used for digitizing. A = anterior; I = inferior; L = left; P = posterior; R = right; S = superior.

there were two perioperative deaths. Of these five patients, four had pulmonary atresia and intact ventricular septum and one had critical pulmonary stenosis. The right ventricular end-diastolic volume was between 5 and 11 ml/m^2 in four patients; two of these underwent a shunt operation as a secondary procedure, and the other two survived with valvotomy alone. Two of the four patients had pulmonary atresia with intact ventricular septum and two had critical pulmonary stenosis. A right ventricular end-diastolic volume greater than 11 ml/m^2 was present in six patients, only one of whom required a subsequent shunt operation in addition to pulmonary valvotomy. As discussed later, this patient had severe residual pulmonary stenosis. Two of the six patients had pulmonary atresia with intact ventricular septum and four had critical pulmonary stenosis. All patients with a right ventricular end-diastolic volume of 5 ml/m^2 or greater survived.

Table 2. Echocardiographic Measurements in 15 Patients

	RV-h (cm)		TV-D (cm)			RV-V (ml/m^2)			
Case	Long Axis	Short Axis	Long Axis	Short Axis	AF (%)	Diastole	Systole	EF (%)	TA-D (cm/m$^{2/3}$)
1	0.7	0.7	0.8	0.9	24.0	19.0	12.3	35	1.4
2	0.7	0.8	1.0	0.8	26.5	5.4	3.3	40	1.5
3	0.6	0.6	1.0	0.9	11.8	16.2	14.0	14	1.6
4	0.4	0.5	0.5	0.3	23.9	4.1	2.5	40	0.7
5	0.6	0.6	0.5	0.5	14.9	2.8	2.1	26	0.8
6	0.6	0.5	1.0	0.9	40.8	13.1	5.2	61	1.5
7	0.6	0.5	1.1	1.0	37.8	20.0	9.7	52	1.8
8	0.5	0.6	0.7	0.8	28.4	8.9	6.1	33	1.3
9	0.5	0.6	1.0	0.9	33.5	6.8	3.4	50	1.6
10	0.6	0.6	1.3	1.3	26.4	11.5	8.1	29	2.1
11	0.6	0.8	0.3	0.3	26.8	1.0	0.7	37	0.5
12	0.6	0.5	0.9	0.7	13.3	18.9	16.2	16	1.4
13	0.6	0.6	0.9	0.8	19.2	10.1	6.9	31	1.4
14	0.7	0.7	0.5	0.5	22.5	3.0	1.9	34	0.9
15	0.7	0.8	0.6	0.5	4.3	3.4	3.2	5	0.9
Mean	0.6	0.6	0.8	0.7	23.6	9.6	6.4	34	1.3
± SD	0.1	0.1	0.3	0.3	9.9	6.6	4.8	15	0.4

AF = area change fraction; EF = ejection fraction; RV-h = right ventricular wall thickness; RV-V = right ventricular volume; TA-D = normalized tricuspid anulus diameter; TV-D = tricuspid valve diameter.

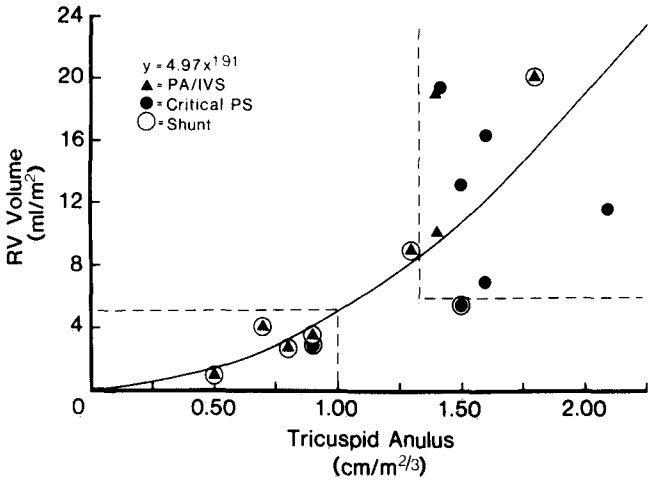

Figure 2. Relation of right ventricular (RV) volume adjusted for body surface area to tricuspid anulus dimension adjusted for the cube root of body surface area in 15 patients with pulmonary atresia and intact ventricular septum (PA/IVS) or critical pulmonary stenosis (PS). The six patients who required shunt procedures are indicated (Shunt). The best fit line **(solid line)** and its equation are shown. The **dotted lines** indicate the region below which a shunt is unquestionably needed (<1.0 cm, <5.0 ml) and the range above which a shunt appears unnecessary (>6.0 ml, >1.4 cm) if adequate relief of right ventricular outflow tract obstruction is provided.

Tricuspid valve dimension. No significant difference was noted for tricuspid valve anulus dimension obtained from long- and short-axis views; for both views it was highly correlated with the right ventricular end-diastolic volume ($r = 0.85$) (Fig. 2). The most significant correlation was obtained using a power function fit. All patients with a right ventricular volume of less than 5 ml/m^2 had a normalized anular dimension of less than 1 cm/m$^{2/3}$. There was broad overlap in ventricular size among patients with an anular dimension of less than 1 cm/m$^{2/3}$. Although an anular dimension of this size was highly predictive of the need for a shunt procedure, it was not possible to define a minimal dimension for which a shunt procedure was unlikely to be necessary.

Ventricular function variables. The diaphragmatic and septal wall thickness measurements did not differ significantly between the two planes (Table 2). There was no significant correlation between wall thickness and right ventricular volume or operative outcome. The area change fraction ranged from 4.3 to 40.8% and was highly correlated with ejection fraction ($r = 0.96$). Neither of these variables was significantly correlated with end-diastolic volume or predictive of either survival or need for shunt procedure.

Combined volume and anulus data. In all subjects with either a normalized right ventricular volume of less than 6.0 ml/m^2 or a normalized tricuspid valve anulus of less than 1.4 cm/m$^{2/3}$, a shunt procedure was needed. In only one patient (Case 7) with either a larger volume or anulus dimension was shunting required. In this patient, inadequate relief of outflow obstruction rather than limited right ventricular size was the apparent reason for intervention.

Discussion

Previous studies. In infants with severe right ventricular outflow tract obstruction, the outcome seems to be dependent on the size of the right ventricle. In 1973, Graham et al. (22) compared different angiographic methods for estimating right ventricular volume, and established regression equations for predicting normal values although only three patients were younger than 4 months of age or had a body surface area of less than 0.3 m^2. They also reported (23) an increase in right ventricular volume after surgery for pulmonary atresia or stenosis with intact ventricular septum in seven patients who underwent combined valvotomy and shunt procedure. In six of these patients, the preoperative right ventricular size was clearly below normal. Absolute ventricular size, therefore, appeared to be a determinant of the type of surgery that should be performed, indicating a need for accurate methods for determining volume measurements in these patients. In an angiographic study (24) of 18 patients with pulmonary atresia and intact ventricular septum, all of whom had a hypertensive right ventricle with reduced cavity size, it was found that inclusion of the outermost borders of the right ventricle in the volume measurement resulted in overestimation of cavity size. Other investigators (18) considered angiographic right ventricular volume determinations in these bizarrely shaped cavities to be of limited value and suggested the use of tricuspid anulus diameter. Because of the close correlation between tricuspid valve diameter and right ventricular size, this more accurately measured variable was thought to provide a better estimation of the true right ventricular size. This measurement alone (25) or in combination (26) with right ventricular dimension has been reported to be predictive of surgical outcome. Very few echocardiographic reports (20,21) exist describing pulmonary atresia with intact ventricular septum. Although it has been reported that the method is reliable for diagnosis, there are few or no data on the usefulness of echocardiographically derived right ventricular volume measurements in these patients.

Echocardiographic results. Our findings indicate that echocardiographic measurement of right ventricular volume and tricuspid anulus diameter is useful in predicting postoperative outcome. Although all patients had decreased right ventricular end-diastolic volume, relief of right ventricular outflow tract obstruction provided adequate palliation in those with a normalized end-diastolic volume greater than 11 ml/m^2. The one patient in this group who required a shunt procedure had severe residual pulmonary stenosis. In contrast, in the group with a normalized right ventricular volume of less than 5 ml/m^2 or tricuspid valve anulus dimension of less than 1 cm/m$^{2/3}$, all required a shunt pro-

cedure. It is unlikely that such a small ventricle can support a normal cardiac index, and adequate pulmonary blood flow should therefore be provided at the initial surgical intervention. Correct management in those patients with intermediate values remains problematic. In our series, it was possible to distinguish those who required a shunt procedure from those in whom only repair of pulmonary stenosis was needed by combined utilization of right ventricular volume and tricuspid valve anulus diameter. Thus, all subjects in this intermediate group in whom a shunt procedure was necessary had either an end-diastolic volume of less than 6.0 ml/m^2 or a valve diameter of less than 1.4 cm/m$^{2/3}$, or both. It must be emphasized, however, that although this provided an exact limit in this patient group, the absolute difference in values between patients on either side of this line (Fig. 2) was minimal. Thus, further data are needed to adequately define this zone of uncertainty.

Methodologic considerations. The accuracy of volume measurements in a minute muscular ventricle is almost certainly quite limited. In addition, normal systolic movement of the heart will introduce a greater degree of error in the calculation of ejection fraction and area change fraction in these patients. It should be noted, however, that the power function relation between tricuspid valve anulus diameter and right ventricular volume resulted in an enhanced ability of this measure to separate patient groups in the low range of values (Fig. 2). The combined use of normalized volume and anular measurements helps overcome the limitations inherent in either method when used alone. Additionally, to the degree that these two variables may function independently, both must be adequate if sufficient pulmonary blood flow is to be maintained, as demonstrated in this study.

Other considerations. The right ventricular size was not related to the wall thickness or ventricular function in patients with severe right ventricular outflow tract obstruction. Echocardiographic calculation of wall thickness, area change fraction and ejection fraction did not provide additional information relevant to the postoperative outcome of these patients. None of these subjects had a giant-sized heart with a Uhl-like (27) ventricle or coronary anomalies. Infants with these lesions appear to receive little benefit from surgical intervention (14). There also appeared to be no inherent difference between subjects with pulmonary atresia and intact ventricular septum and those with critical pulmonary stenosis. For both groups it was the right ventricular and tricuspid valve anulus size that predicted outcome, rather than the presence or absence of valve atresia.

Implications. Echocardiographic determination of right ventricular end-diastolic volume and tricuspid anulus diameter provides an excellent means of predicting the postoperative outcome in patients with reduced right ventricular size. This method should prove useful in the preoperative selection of optimal surgical therapy.

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